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Combination therapy using PSE and TIO ameliorates hepatic encephalopathy due to intrahepatic portosystemic venous shunt in idiopathic portal hypertension

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Abstract

A 64-year-old woman treated for anemia and ascites exhibited hepatic encephalopathy. Abdominal ultrasonography and computed tomography (CT) showed communication between the portal vein and the middle hepatic vein, indicating an intrahepatic portosystemic venous shunt (PSS). Since hepatic encephalopathy of the patient was resistant to medical treatment, interventional radiology was performed for the treatment of shunt obliteration. Hepatic venography showed anastomosis between the hepatic vein branches, supporting the diagnosis of idiopathic portal hypertension (IPH). To minimize the increase in portal vein pressure after shunt obliteration, partial splenic artery embolization (PSE) was first performed to reduce portal vein blood flow. Transileocolic venous obliteration (TIO) was then performed, and intrahepatic PSS was successfully obliterated using coils with *n*-butyl-2-cyanoacrylate (NBCA). In the present case, hepatic encephalopathy due to intrahepatic PSS in the patient with IPH was successfully treated by combination therapy using PSE and TIO.

Keywords

Intrahepatic portosystemic venous shunt (PSS), hepatic encephalopathy, transileocolic venous obliteration (TIO), partial splenic artery embolization (PSE), idiopathic portal hypertension (IPH)

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Introduction

Intrahepatic portosystemic venous shunt (PSS), which is communication between the intrahepatic portal veins and the systemic veins, has rarely been reported since the first report by Raskin et al. in 1964 (1). Intrahepatic PSS is now encountered more frequently with the developments of diagnostic imaging (2,3). Clinical manifestations of intrahepatic PSS depend on the shunt flow, and a shunt with a high flow may cause hepatic encephalopathy. In this condition, portal blood flows directly into the systemic circulation without passing through the liver, causing hepatic encephalopathy (4). The location of PSS is extrahepatic in most cases, but a few cases of intrahepatic PSS have been reported (5,6). This report describes a case of idiopathic portal hypertension (IPH) whose hepatic encephalopathy was successfully treated by combination therapy using partial splenic artery embolization (PSE) and transileocolic venous obliteration (TIO).

Case report

A 64-year-old woman presented with notable leg edema. As cardiomegaly and marked anemia were recognized, this patient was referred to our hospital

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for further examinations. The liver was not palpable and three finger-widths of spleen were palpable. Abdominal fluctuation was present. Pronounced edema was seen in the lower extremities. No history of prescription drugs nor alcohol was reported. Informed consent for the case study was obtained.

Laboratory data were as follows: white blood cell count, 1500/μL; hemoglobin (Hb), 2.1 g/dL; platelet count, 8.2×10^4/μL; prothrombin time, 35% (normal range, 70–140%); albumin, 2.9 g/dL (normal range, 4.1–5.0 g/dL), and brain natriuretic peptide, 626.6 pg/mL (normal range, 0–18.4 pg/mL), suggesting marked anemia and pancytopenia, impaired hepatic function, and cardiac failure.

Ultrasonographic examination of the abdomen showed cirrhosis-like findings, such as rounded hepatic margins, a slightly irregular liver surface, and mild parenchymal coarseness. Communication between the portal vein and the middle hepatic vein was visualized in segment 8, indicating intrahepatic PSS. Enhanced computed tomography (CT) revealed marked splenomegaly, massive ascites, and collateral blood vessels near the abdominal wall. Early visualization of the contrast medium in the hepatic veins was recognized, suggesting the existence of intrahepatic PSS. Three-dimensional (3D) CT at the portal phase after injection of contrast medium revealed intrahepatic PSS between the portal vein branch and the middle hepatic vein (Fig. 1).

Angiography of the celiac and superior mesenteric arteries was performed. However, identification of the shunt location was difficult because of high blood flow in the shunt. Hepatic venous wedge pressure measured from the right hepatic vein was not increased (10 mmHg). Anastomosis between the hepatic vein branches was recognized by hepatic venography (Fig. 2).

Liver biopsy specimens showed that the basic structure of the hepatic lobules was preserved. Regenerating nodules were not recognized. Portal veins were indistinct or invisible in the portal areas. In some portal areas, the shape of the portal vein was irregular, suggesting an aberrant vessel. These pathological findings are consistent with IPH.

Edema of the extremities and ascites resolved with protein intake restriction, salt intake reduction, and diuretic administration. However, hepatic encephalopathy manifested and was difficult to control even with administration of lactulose and branched chain amino acids. The hepatic encephalopathy was thought to have resulted from portal flow steal through the intrahepatic PSS. Occlusion of the intrahepatic PSS was planned for the treatment of hepatic encephalopathy, but there was concern that occlusion of the shunt may result in an increase in portal vein pressure. Therefore, PSE was first performed using coils and gelfoam to reduce portal blood flow towards the liver. Approximately 70% splenic infarction was achieved after PSE (Fig. 3). TIO was performed 6 weeks later when the effects of PSE on the spleen disappeared, judging from both clinical point of view and enhanced CT findings. The distal ileum was exposed under a small abdominal incision. A catheter was advanced into the portal venous system via the ileocolic vein. Intrahepatic PSS was obliterated by using a total of 20 coils of various types, including detachable coils. Since the blood flow of intrahepatic PSS persisted, 4 mL of *n*-butyl-2-cyanoacrylate (NBCA)/lipiodol (1:1) was injected to the intrahepatic PSS, resulting in complete occlusion of the shunt (Fig. 4). Enhanced CT scan showed complete occlusion of the intrahepatic PSS and the presence of thrombus up to the inflow of the portal vein.

The changes in portal blood flow measured by Doppler ultrasonography, portal vein pressure, and

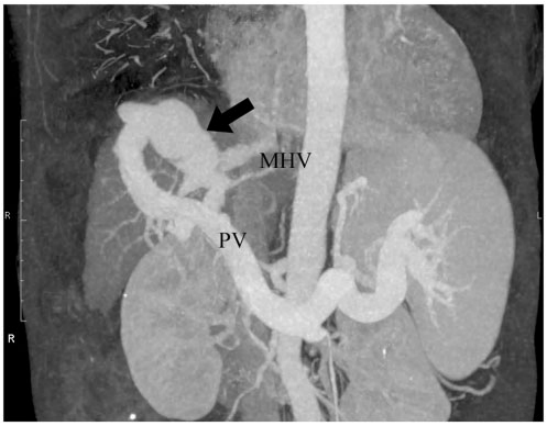

Fig. 1. 3D CT at the portal phase. 3D-CT revealed intrahepatic PSS (arrow) between the portal vein branch (PV) and the middle hepatic vein (MHV).

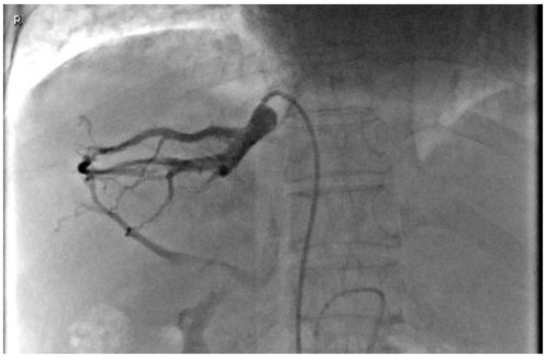

Fig. 2. Hepatic venography. Hepatic venography showed anastomoses between the hepatic veins.

ammonia from before to after PSE and TIO are shown in Table 1. The blood flow of the portal vein and the splenic vein before PSE was 2980 mL/min and 2820 mL/min, respectively, and it was reduced to 1482 mL/min and 1378 mL/min, respectively, after PSE. After TIO, the blood flow in these vessels slightly increased to 1740 mL/min and 1440 mL/min, respectively. The flow rate in the middle hepatic vein clearly decreased from 133 cm/s to 81 cm/s after TIO. Hepatic venous wedge pressure (HVWP) was normal (10 mmHg) before PSE. Portal vein pressure increased from 14 mmHg to 24 mmHg after TIO, increasing the status of portal hypertension. After TIO, ammonia levels increased from 203 μg/dL to 243 μg/dL in the portal vein and decreased from 115 μg/dL to 77 μg/dL in the hepatic vein. The difference in the ammonia levels

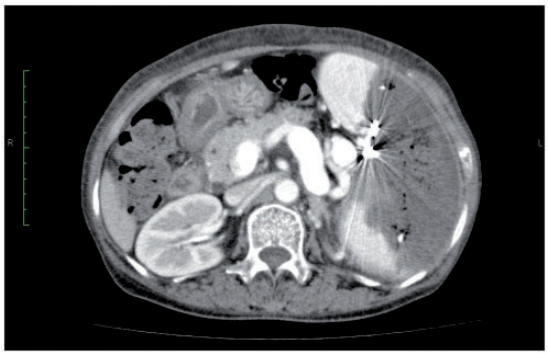

Fig. 3. Enhanced CT at the arterial phase after partial splenic artery embolization (PSE). A low density area was found in the spleen after PSE. The infarction rate was approximately 70%.

Table 1. Changes in portal blood flow, portal vein pressure, and ammonia levels pre and post PSE and post TIO.

	Pre PSE	Post PSE	Post TIO
Blood flow (portal vein) (mL/min)	2980	1482 ↓	1740 ↑
Blood flow (splenic vein) (mL/min)	2820	1378 ↓	1440 ↑
Flow rate (middle hepatic vein) (cm/s)	125	133 ↑	81 ↓
Portal vein pressure (mmHg)		14	24 ↑
NH_3 (portal vein) (μg/dL)		203	243 ↑
NH_3 (hepatic vein) (μg/dL)		115	77 ↓
ΔNH_3 (μg/dL)		88	166 ↑

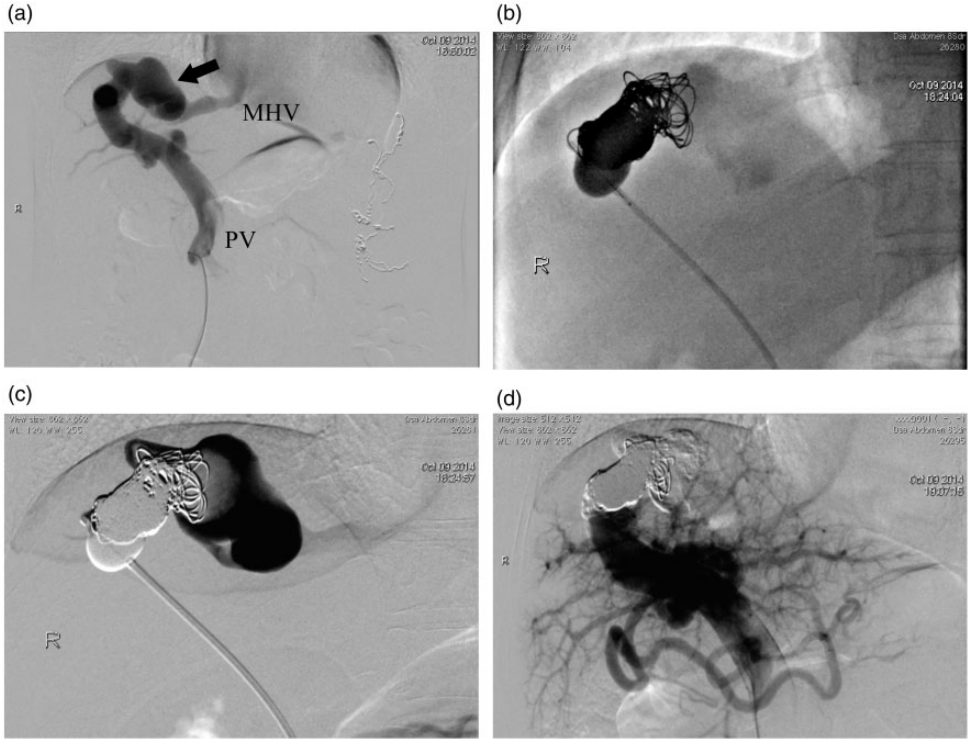

Fig. 4. Transileocolic venous obliteration (TIO). (a) Pre TIO. Intrahepatic portosystemic venous shunt (PSS) indicated by an arrow was found between the portal vein branches (PV) and the middle hepatic vein (MHV). (b) Post coiling. Several coils were placed in the intrahepatic PSS. (c) Prior to NBCA. N-butyl-2-cyanoacrylate (NBCA) was additionally administered into the intrahepatic PSS. (d) Portography after TIO. Intrahepatic PSS were completely occluded and the portal vein branches were visualized after TIO.

between the portal vein and the hepatic vein showed an increase from 88 μg/dL to 166 μg/dL, indicating a nearly twofold improvement in ammonia clearance. An obvious decrease in ammonia levels after obliteration of intrahepatic PSS resulted in clinical improvement of hepatic encephalopathy.

On admission, esophageal varices were classified as Lm, F2, Cb, RC1 with hematocystic spots according to the general rules for recording endoscopic findings of esophagogastric varices (7). Esophageal varices were immediately treated with endoscopic variceal ligation (EVL). However, as a result of portal hypertension caused by TIO, esophageal varices worsened to Li, F2, Cb, RC2, and Lg (-), and they were again treated with EVL. The brain function was resolved after treatment and the patient is currently attending our hospital without sequelae.

Discussion

The portosystemic shunt is extrahepatic in most cases and intrahepatic PSS is extremely rare. Park et al. classified 14 cases of intrahepatic PSS in the literature into four morphologic categories (8). In this case, ultrasonography, CT, and angiography revealed communication between the portal vein and the middle hepatic vein, corresponding to Park's type 2. Type 2 intrahepatic PSS is a localized peripheral shunt in which single or multiple communications are found between peripheral branches of the portal and hepatic veins in one hepatic segment. In the aneurysmal type, a portal aneurysm may precede the venous shunt and then rupture into the hepatic vein, resulting in the formation of communication (9). Remer et al. also reported that most cases of intrahepatic PSS were located in the left lobe and had aneurysmal communication between the portal and hepatic veins (10).

Much remains unclear regarding the etiology of intrahepatic PSS and the stage of its formation. Two theories, i.e. the congenital theory and the acquired theory, have been postulated (1,11,12). The former suggests a persistent embryonic venous anastomosis while the latter suggests that the shunt results from portal hypertension, trauma, or rupture of a portal vein aneurysm. Kozuka et al. reported that microscopic findings of patients with intrahepatic PSS showed both the muscular layer and the elastic lamellae disappeared abruptly from the wall of the shunt, and cerebral manifestation was not apparent until older age (13). These results suggest that intrahepatic PSS is acquired. In the present case, it was speculated that acquired anastomosis had formed between the middle hepatic vein and intrahepatic portal branch as a result of vascular malposition as well as portal hypertension recognized in IPH.

Treatments of PSS are required for patients with hepatic encephalopathy. Surgical closure of the shunt was previously used to alleviate symptoms. Recently, less-invasive treatments using interventional radiology (5,14,15), such as balloon-occluded retrograde transvenous obliteration (B-RTO), percutaneous transhepatic obliteration (PTO) and TIO, have been performed to obliterate PSS. B-RTO is a relatively non-invasive technique and is recommended as the first choice for treatment of portosystemic venous shunt (14,15). In this case, a catheter could not be placed at the site of PSS because of its high blood flow, so B-RTO approached from the middle hepatic vein was considered impossible. Although PTO was initially considered, the sites of paracentesis and occlusion were in proximity to each other, which may have complicated catheter manipulation. Therefore, TIO was chosen to occlude the intrahepatic PSS. In the treatment of intrahepatic PSS, increased portal vein pressure should be taken into account after shunt obliteration. Anticipating that portal hypertension would be exacerbated after shunt obliteration, PSE was performed first to reduce the portal blood flow before shunt obliteration. In this way blood flow in the main portal vein was reduced by almost 50%. Despite having performed PSE, portal pressure increased by approximately 70% after TIO.

In conclusion, we have reported a rare case of hepatic encephalopathy due to intrahepatic PSS in a patient with IPH. The intrahepatic PSS was successfully treated by combination therapy using PSE and TIO, resulting in marked improvement of shunt encephalopathy.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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